

Extraordinary manifestations of chronic pyonephrosis¹

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Summary: Twenty-five patients who underwent nephrectomy for chronic pyonephrosis were studied retrospectively. Such patients may present with a wide range of symptoms. Marked haematological and biochemical abnormalities are found, the most striking being a gross elevation of plasma viscosity (or ESR) and a raised alkaline phosphatase. The resulting picture often suggests extrarenal disease, and diagnostic confusion occurred in a number of patients.

Introduction

Although a pyonephrosis would be expected to cause loin pain and symptoms of urinary tract infection, increasingly we have found that chronic cases are referred not directly to a urologist but via a physician or other specialist to whom the patient has presented with nonspecific and often confusing symptoms (Kirk 1982). The present retrospective study of a group of patients who underwent nephrectomy for a destructive chronic pyonephrosis emphasizes those features, unexpected in urinary tract disease, which might cause such confusion.

Patients

The case records of 25 patients were studied retrospectively. There were 21 women and 4 men. Their ages ranged from 32 to 78 years. Eighteen had staghorn or multiple renal calculi, 2 an obstructing ureteric calculus, and 2 a primary pelvic hydronephrosis. One patient had previously had a matrix calculus removed, and in 2 the cause of the non-functioning inflamed kidney was uncertain. In all cases an intravenous urogram (IVU) had demonstrated a non-functioning kidney, which proved on removal to have been destroyed by chronic inflammation, often with associated xanthogranulomatous pyelonephritis (see below).

Results

Presenting symptoms: Symptoms recorded in the case notes are listed in Table 1. The terms 'loin pain' and 'abdominal pain' seek to distinguish pain suggesting renal pathology from that of a nonspecific or atypical distribution. The patient with pain in her legs had this as a sole complaint and initially had been referred to a rheumatologist. Another woman's swollen leg proved to be due to the physical effect of a retroperitoneal swelling and not the deep vein thrombosis originally diagnosed. It is of note that in only 4 patients' notes was there recorded a past or present history of symptomatic lower urinary tract infection.

Investigation: Twenty-two patients had a significant pyuria. On culture a significant pure growth was obtained in 11 and a mixed growth in 3. Available haematological and biochemical results of interest are shown in Figures 1 and 2.

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Table 1. Presenting symptoms in 25 patients with chronic pyonephrosis

Loin pain	10	Anorexia	10
Abdominal pain	9	Weight loss	15
Nausea/vomiting	10	Tiredness	9
Diarrhoea	4	Pain in legs	1
Constipation	1	Calf swelling	1

Table 2. Non-contributory investigations resulting from confusing presentation

Barium meal	4
Barium enema	4
Cholecystogram	2
Bone marrow aspiration	4
CAT scan and/or ultrasound●	4
Spinal X-rays	1

● Excluding those performed after an IVU

Although ultimately all patients underwent IVU, in many cases this was preceded by investigations directed towards other systems (Table 2). Indeed, it was the incidental finding of a staghorn calculus or enlarged kidney during these investigations in a number of cases which pointed to the true diagnosis.

Diagnosis: As a result of the variety of presenting symptoms and the findings on initial investigation (Figures 1 and 2), considerable diagnostic difficulty was encountered with a number of these patients. In only 12 was their illness immediately recognized as being due to renal disease. Eight patients, who have been reported in more detail elsewhere (Kirk 1982), had illnesses characterized by marked weight loss and debility and were thought to be suffering from some form of malignancy. Three appeared to have primary gastrointestinal disease, one man's enlarged kidney was thought to represent splenomegaly, and the woman with painful legs was considered to have an arthropathy.

Treatment: As indicated, all patients underwent nephrectomy. In 2 this was preceded by a short period of drainage via a percutaneous nephrostomy. Another had a perinephric abscess drained, but it was only after a nephrectomy some months later that her anaemia resolved and her elevated alkaline phosphatase and viscosity returned to normal. Following nephrectomy, all patients ultimately returned to good health with reversal of the haematological and biochemical abnormalities.

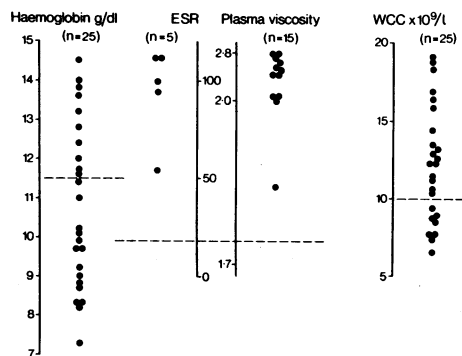


Figure 1. Haematological findings available from retrospective review of the records of 25 patients with chronic pyonephrosis. (Note: patients were treated in three different hospitals, hence some had ESR rather than plasma viscosity recorded. ESR is indicated on a linear scale, and the viscosity scale arranged so that points on it have similar significance to points at same level on ESR scale)

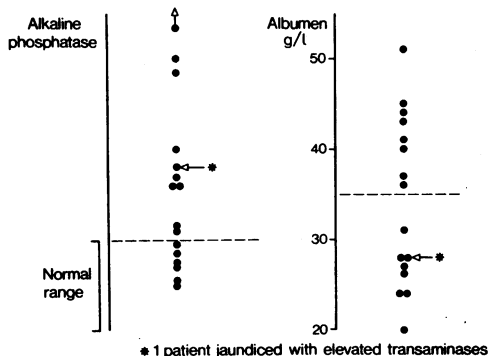


Figure 2. Biochemical findings available from records of 25 patients. (Note: different laboratories used different units to record level of alkaline phosphatase. Points on chart recorded in proportion to the normal range for the laboratory reporting the test)

Pathology: All the removed kidneys showed extensive chronic or acute-on-chronic inflammatory changes. In all but 4 there were areas of xanthogranulomatous pyelonephritis, although this was extensive, involving most of the kidney, in only 9 of the kidneys.

Discussion

This paper describes an unselected group of patients who were not specifically chosen because of atypical features. Although many presented with loin pain and other features which enabled prompt recognition of the diagnosis, some caused considerable confusion. Even where there were clear indications of renal disease, it was noted that many of the non-urological symptoms listed in Table 1 were also present, and on investigation the abnormalities shown in Figures 1 and 2 often were found. Thus the presence of a chronic pyonephrosis seems to produce a definite syndrome. Patients present with weight loss, tiredness, gastrointestinal symptoms, with or without loin pain, and on investigation are often anaemic, invariably have a grossly elevated ESR/plasma viscosity, and about half have an elevated alkaline phosphatase and depressed serum albumin. A pyuria is usual but about half will have a sterile urine. When the non-urological manifestations predominate, it is not surprising that diagnostic confusion can arise.

The investigations which produced most difficulty in some of the patients were the ESR/viscosity and the elevated alkaline phosphatase. The former was so grossly raised, to a level associated with conditions such as myelomatosis, that this finding alone seemed to confirm an initial impression of malignant disease (Kirk 1982). Although not characterized in most cases, in the 4 for which the information is available the raised alkaline phosphatase was of hepatic origin. Although it is appreciated that a reduced serum albumin is a nonspecific abnormality not especially attributable to hepatic disease, its association with a raised alkaline phosphatase suggests 'abnormal liver function tests', with all that that implies – and that is not usually taken to be renal disease.

Stauffer (1961) described a patient with renal carcinoma who had hepatosplenomegaly associated with bromsulphthalein retention, elevated alkaline phosphatase and protein abnormalities in the absence of liver metastases. This condition of nephrogenic hepatic dysfunction subsequently was described in two patients with xanthogranulomatous pyelonephritis (Vermillion *et al.* 1970). The latter occurs as a result of chronic inflammation, usually in association with stone disease and/or obstruction, and is characterized histologically by granuloma formation and the presence of 'foamy xanthomatous histiocytes' (Gingell *et al.* 1973). Such changes were present in most but not all of the patients' kidneys in our series. However, it was not possible to correlate the 'hepatic' abnormalities with this condition and in one of the 4 in whom xanthogranulomatous changes were absent, there was an elevated alkaline phosphatase. It is likely that these abnormalities represent a hepatic response to extrahepatic sepsis, which has been described with disease in various organs (Neale *et al.* 1966). The xanthogranulomatous changes are a reflection of the severity of the sepsis producing the hepatic disorder rather than its direct cause.

All these patients returned to good health after nephrectomy. However, this good prognosis is put in jeopardy if the diagnosis is delayed. This condition should be considered in patients with debilitating disease of uncertain origin, and in such patients the finding of a urinary infection or a pyuria must be taken seriously. The surgeon operating on a patient with a chronic pyonephrosis should be aware that many of these patients have a marked systemic illness and require careful pre- and postoperative care.

References

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